Journal of the Saudi Heart Association (2010) **22**, 179–185

King Saud University

Journal of the Saudi Heart Association

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EDITORIAL

Three dimensional imaging using 64 detector row multi-slice CT should be used more widely for the diagnosis and management of congenital heart disease

Although 64 detector row multi-slice CT (MSCT) is the latest advance in multi-slice technology, it remains less frequently utilized for the diagnosis and management of congenital heart disease (CHD) compared to diagnostic cardiac catheterization (DCC) with biplane angiography, two dimensional echocardiography (2DE), and magnetic resonance imaging (MRI). When used, the interpreting physician is usually a radiologist who visualizes 2D slices in the axial plane and rarely in three dimensions (3D). Many pediatric cardiologists complain that the radiation exposure (RE) from MSCT is too high to ethically justify its use in children, but ignore the effective dose to which their patients are exposed during DCC and interventional CC (ICC) using biplane fluoroscopy and cineangiography. Gating image acquisition to the ECG signal is rarely done because it is thought to dramatically increase RE. And the literature is not very helpful. Methods to calculate RE are complex especially in infants and children, arcane, and very poorly explained. Radiation units of measure and their meaning vary from report to report, and explanatory language is abstruse and often incomprehensible to those who would order such tests. The natural history of many complex forms of congenital heart disease is incompletely understood in individuals who might benefit from a detailed anatomic 3D analysis especially for those lesions that are obligatorily palliated and thus have a different natural history and risk of developing radiation sequelae. Amid this confusion, the default imaging methods for CHD are 2DE, DCC, and MRI, because 2DE and MRI entail no radiation, and DCC has been used for more than half a century with RE that is universally accepted as safe.

Appropriately, 2DE is the primary imaging modality for congenital heart disease (CHD) at all ages. Pediatric cardiologists stress anatomy while their adult counterparts emphasize physiology. When physiologic measurements are sought in infants and children, they are often acquired with the patient crying or struggling to escape, thus obviating their value. This is especially true with sub-costal imaging which most unsedated infants and children resist. A common 2DE acquisition format consists of sweeps, moving the transducer from location x to location y during image acquisition in an attempt to show the relationship of structure a to structure b. After the fact, these phrenetic images are incomprehensible except to the person who acquired them. Unfortunately, infants and children are rarely routinely sedated, and when they are, a variety of drugs is used in dosages that often are too small to allow a comprehensive examination. And a full understanding of how to sequentially acquire a set of 2D black white, color flow, and Doppler images that represent a 3D whole is rarely taught or understood. These self-imposed pediatric 2DE limitations cry out for improvements in techniques that allow better measurement of pressures, flows and resistances. Ironically adult echocardiographers have been using 2DE for this purpose for years, but in pediatrics this information is still sought primarily through DCC.

Everyone accepts that patients need anesthesia for DCC and ICC in order to eliminate pain and prevent movement. Many forget that the primary need is to obtain a representative physiologic steady state during which pressures and oxygen saturations can be accurately obtained and flows and resistances calculated. The corresponding need for infants and children to be sedated during 2DE seems intuitive, but is rarely effectively practiced. And ignored are the depressant effects of general anesthesia on cardiac function (Filner and Karliner, 1976), the most common sedation modality for both DCC and ICC. Cardiac output drops, and the data often bear little resemblance to the awake steady state. Witness the child with an aortic systolic pressure of 80 mmHg under general anesthesia whose pressure when awake or under light sedation is 110! Nevertheless, the numbers obtained are accepted as accurate making the term "DCC" highly problematic.

Unfortunately today pediatric and adult cardiac trainees are more likely to be exposed to and taught how to do interventional procedures rather than to understand and record

doi:10.1016/j.jsha.2010.07.005

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accurate steady state physiology. And it is now rare to calculate LV or RV volumes, ejection fractions, and flows angiographically. These values are simply "eyeballed" from whatever imaging format is used.

Even though DCC is well established as the standard for imaging CHD, its radiographic application varies from center to center, the technology is old, anatomy is poorly or incompletely visualized, and it requires use of large volumes of contrast agents with their attendant risk of allergic reactions and renal functional impairment. Each angiogram requires a separate bolus. And angiography "sees" only that pathology within the one or two 2D orthogonal planes visualized and only to the degree there is adequate contrast.

As the number of 2DE increases, the number of DCC is falling, but the need to see anatomy accurately and record physiology faithfully remains unchanged. To meet this need new imaging modalities are emerging including, MSCT, 3D echo (3DE), and MRI.

Cardiologists commonly hold that MRI represents the gold standard for ventricular volumetric analysis and that these calculations are automatically rendered by the application of MRI cardiac software. For example, operated patients with tetralogy of Fallot with chronic moderate or severe pulmonary regurgitation by 2DE often undergo MRI in order to obtain RV end-diastolic volume as an index to help determine when to replace the pulmonary valve (Lindsey et al., 2010). And while these data have long been available using standard biplane angiography or more recently by 3DE (Hubka et al., 2002), few laboratories retain the expertise or have the necessary equipment. What is not so well known is that, like 2DE and angiography, ventricular shapes by MRI must be accurately drawn by hand for true volumes to be calculated. Failure to understand the differences between LV and RV morphology with different CHD and physiologic conditions or failure to accurately identify end-systole and end-diastole will produce significant errors. Few physicians realize that volumes and outputs can be calculated for both ventricles and atria using MSCT.

Although MRI offers the potential for 3D imaging with no radiation risk, albeit with a lower spatial resolution compared to MSCT (Friedrich, 2010), few radiologists bother to reconstruct MRI in 3D. MRI requires longer anesthetic times averaging 30 min to an hour or longer and is not as easily scheduled, and fewer centers have access to expensive modern MRI cardiac software. Compared to MSCT, MRI is no less subject to errors and is simply not as practicable.

1. What is MSCT?

64 MSCT uses an X-ray beam that rotates around the patient approximately three times per second with the patient moving through the beam (called pitch) to cover the field of view and expose charged coupled devices that create 64 2D slices with a thickness as thin as .6 mm. Excellent spatial resolution results. Approximately 3.8 cm of depth is scanned with each revolution, making data acquisition remarkably fast. No matter what image acquisition algorithm is used, the entire field of interest can be scanned in less than 15 s and usually in less than 10. Although radiologists chose for expediency to view only 2D slices, computer rendering can easily stack these slices to produce a 3D volumetric rendering. 2D and color 3D images can be displayed in any plane, and the entire volume can be rotated around a single point.

Images are acquired in any one of three acquisition algorithms: non-ECG gating and prospective or retrospective ECG gating. With non-gating, the X-ray beam is activated only during a random single phase within the cardiac cycle. For prospective gating, a single specified time within the cycle is empirically chosen, usually 75% of the R–R interval for heart rates under 80 bpm and 55% for rates greater than 80. These times are associated with the least cardiac motion. With non-gating and prospective gating, RE is least. With retrospective gating, the entire cardiac cycle is acquired necessitating exposure over multiple heart beats that results in RE that can be multiples of that achieved during non-gating or prospective gating algorithms. It is these high doses that have been publicized in the lay press that have contributed to fear of CT radiation exposure. Research is underway to produce algorithms that activate the beam only at end-diastole and end-systole rather than the entire cardiac cycle, thus substantially reducing RE compared to retrospective gating. Of course the lower the kV and mAs, the lower the RE.

2. What is the RE during MSCT compared to DCC, and is MSCT unethical?

Bacher et al. reported median RE during standard pediatric biplane DCC and ICC at 4.5 and 6 mSv, respectively (Bacher et al., 2005). These exposures are universally accepted as safe. Nevertheless, this is roughly equivalent to the natural human background RE over the course of one to two years in Sunbelt regions of the world. The RE for cardiac CT in children is difficult to determine and understand, but current estimates suggest that for retrospectively gated CT with kV and mAs set to lowest practical values, RE is in the range of 10–15 mSv (Hollingsworth et al., 2007), roughly 3–4 times the published median values for DCC and ICC.

Such data tell only part of the story and may be misleading. The actual RE from DCC and ICC varies widely because the use of fluoroscopy is operator dependent and uncontrolled, most of the RE (50%) occurs during angiography which is directly proportional to the length of the cine runs (Rassow et al., 2000) and is operator dependent, and some procedures last for hours. Few operators are disciplined in the control of the fluoroscopy foot pedal. They simply keep the fluoro activated for the time they need to see the relationship between catheter and cardiac silhouette sometimes resulting in almost continuous radiation exposure. Legal statutes require only that fluoroscopy time be recorded, even though that time is never translated into effective RE. In short, the true RE for infants and children during DCC and ICC is completely unknown and likely much greater than is assumed or published. Rassow et al. (2000) reported effective RE in infants during DCC to range from 2 mSv (25th percentile) to nearly 18 mSv (90th percentile), a range that includes non-gated and prospectively gated CT studies at the low end and retrospectively gated CT at the other (Frush and Yoshizumi, 2006). Unlike DCC and ICC, when radiation exposure is set for MSCT, it is fixed and controlled by the application software, not the operator.

Given the limitations in determining RE in infants and children and the uncontrolled RE during cine and fluoroscopy, the assumption that cardiac CT is unethical on the grounds that it

exposes children to too much radiation is simplistic and unwarranted. As with DCC and ICC, Cardiac CT has risks associated with radiation that must be balanced against the value of the information obtained relative to the risk which is partly a function of the expected natural life span of the patient. Patients with the most complex lesions such as single ventricle have shorter life spans than those with less complex lesions and therefore have a different risk/benefit equation. And their anatomy and physiology often defy easy analysis by any imaging modality. Relative to DCC and its attendant RE, MSCT has the potential to provide better and more comprehensive anatomic imaging especially when rendered in 3D.

3. Why MSCT?

MSCT allows one to display individual parts as 3D images either singly or combined as a whole. 3D anatomy is more intuitive than either biplane 2D standard angiography or 2D CT or MRI and is preferred by most surgeons and non-imaging cardiologists because they can interpret 3D images independently. With 2D imaging, an intervening expert is needed to explain the images especially when they are shown to an audience. This is especially true when sweeps are acquired. In these settings, surgeons often demand another imaging modality that more easily and completely meets their needs. One has only to observe which images conference participants persistently attend to when both 2DE and MSCT 3D images are displayed simultaneously.

As a cardiologist I prefer to render all parts in 3D including the LV and aorta, RV and pulmonary arteries, RA and systemic veins, LA and pulmonary veins, coronary arteries, and the airway. I then display them in contrasting colors either as an integrated whole with or without partial transparency or as groups of parts in a rendering that helps to explain the anatomy. Fig. 1 shows tetralogy of Fallot in an infant with pulmonary atresia where individual parts have been combined into a whole. The 2DE suggested a double aortic arch. MSCT revealed a ''U'' shaped ductus from the base of the innominate bifurcation which fed the left pulmonary artery. The true arch was on the right, and a vascular ring was not present. Unlike 2DE, the 3D anatomy was not ambiguous.

Fig. 2 shows a non-gated 3D rendering of only the right and left atria and pulmonary veins. In this rear projection we see the SVC and RA in natural colors and the LA and pulmonary veins in violet. Note the complete isolation of the right upper and middle pulmonary veins from the left atrium.

Non-gating, like prospective gating, subjects patients to the least RE and is best for visualizing non-moving structures such as the great arteries and veins and the airway. Fig. 3 shows a non-gated aortic isthmus after balloon angioplasty. Some would argue that the result is satisfactory. However, using another important feature of MSCT as shown in Fig. 4, the same arch has been straightened and rotated about its long axis to show the isthmus and an anterior shelf in relief. A persistent obstruction is obvious (yellow arrow). The coronaries also happened to be well seen.

Prospective and retrospective gating, by allowing one to choose those images where movement is least, greatly improve spatial resolution allowing one to routinely see the coronary

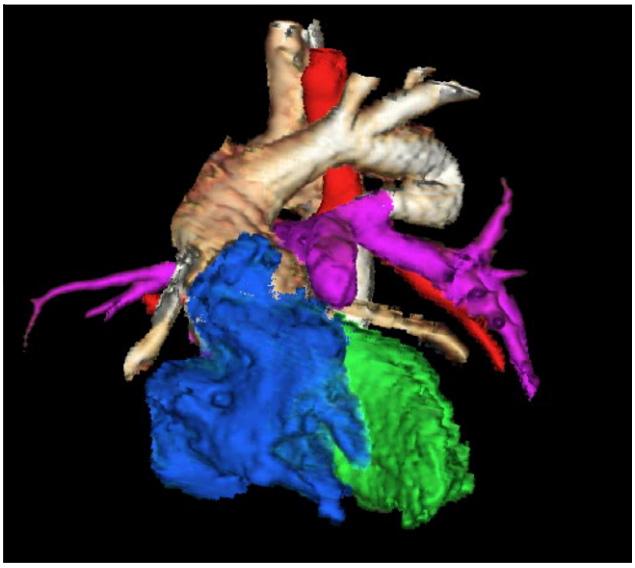

Figure 1 RV (blue); LV (green); Pas (violet); trachea (red).

Figure 2 Posterior view of left atrium and pulmonary veins (violet). Note complete isolation of the right upper and middle pulmonary veins.

arteries or at least their origins. For most decision making in CHD, one rarely needs to see the secondary coronary arteries or the full length of the major branches. Fig. 5 shows an example of a teenager with exercise-induced syncope who was found to have complete absence of the left main coronary artery. Fig. 6 is a patient with Kawasaki disease with giant calcified sacular aneurysms.

Retrospective gating also allows one to measure end-systolic and diastolic volumes and calculate ejection fractions, stroke volumes and Qp/Qs ratios, and pick that phase of the

Figure 3 Left anterior view of the aortic arch, brachiocephalic vessels, and coronary arteries post-balloon angioplasty for coarctation. Appearance suggests satisfactory result.

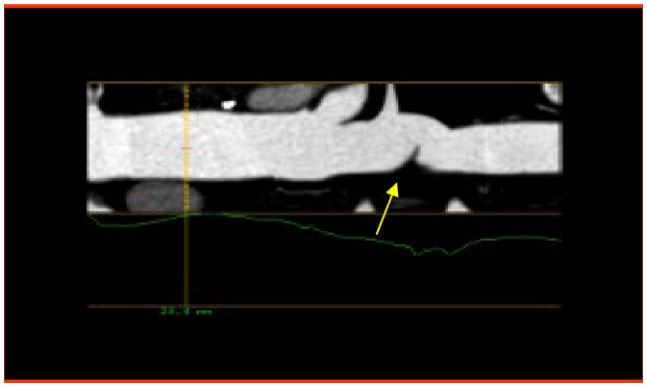

Figure 4 Same arch shown in Fig. 3 straightened and rotated around its long axis to reveal a protruding anterior shelf (yellow arrow) that narrows the internal lumen by approximately 50%.

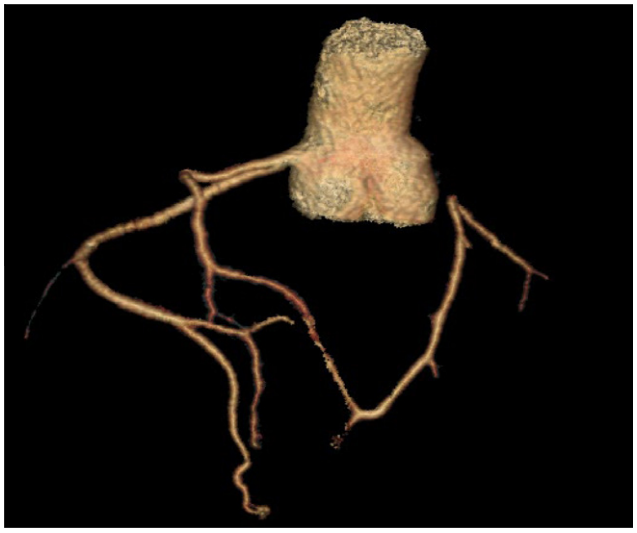

Figure 5 Aortic root in a teenager with syncope. Note absence of the left main coronary artery. The left anterior descending coronary is supplied by the conus branch of the right coronary artery.

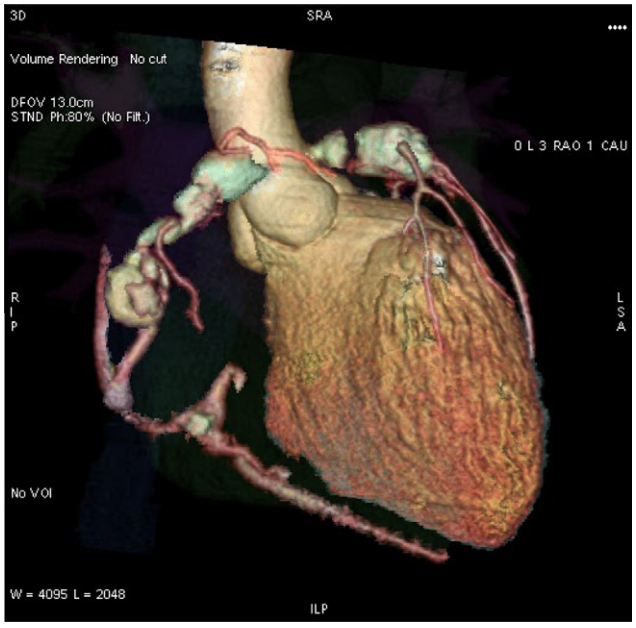

Figure 6 Right and left coronary artery giant saccular aneurysms in a child with Kawasaki disease.

cardiac cycle where motion artifact is least. Fig. 7 shows end-systolic and diastolic frames from the left and right ventricles in a patient with a large VSD. RA and LA volumes can also be calculated. These measurements and images add important physiologic insights that help one to understand the lesion more completely and assist in decision making. For example, a ventricular septum convex toward the RV indicates sub-systemic peak pressures. Gross enlargement of the left or right atrium suggests significant AV valve regurgitation and/or elevations in mean atrial pressure.

The airway can only be inferred from DCC and 2DE, but with MSCT it and the esophagus are easily rendered. One can also create virtual bronchoscopy or arterioscopy. Fig. 8 shows a rear view of an infant with tetralogy of Fallot with absent pulmonary valve in which the huge right and left branch pulmonary arteries completely occlude both the right and left main stem bronchi (shown in red). Fig. 9 shows arterioscopy of the distal aortic arch of an infant with coarctation of the aorta, a narrow isthmus, and a large ductus arteriosus.

For pulmonary sling, vascular rings, and double aortic arch, MSCT is the only test needed since it shows both the offending arterial vasculature and the encircled airway and esophagus. Fig. 10 shows a right arch and a vascular ring with an associated diverticulum of Kumerol. The ring consists of the arch, diverticulum, branch pulmonary arteries, and the invisible ligamentum between the distal diverticulum and the left pulmonary artery.

CT image acquisition is fast even with retrospective gating, taking less than 10–15 s. Because the preparation, acquisition, and turnaround times combined are often less than 30 min, CT is easily scheduled anytime. Unlike MRI, scanning patients on ECMO or with implanted pacemakers or other metallic hardware is not a contraindication. Intravascular stents are nicely rendered as shown in Fig. 11. Even neo-intima formation is readily apparent as shown in Fig. 12.

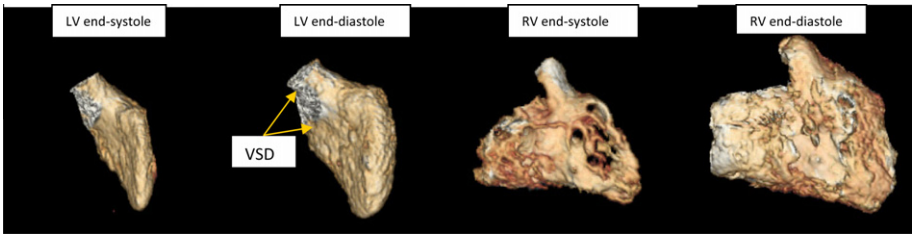

Figure 7 RV and LV end-diastolic and end-systolic images in a patient with large VSD (yellow arrows) used to calculate RV and LV end-diastolic and end-systolic volumes, ejection fractions, and stroke volumes.

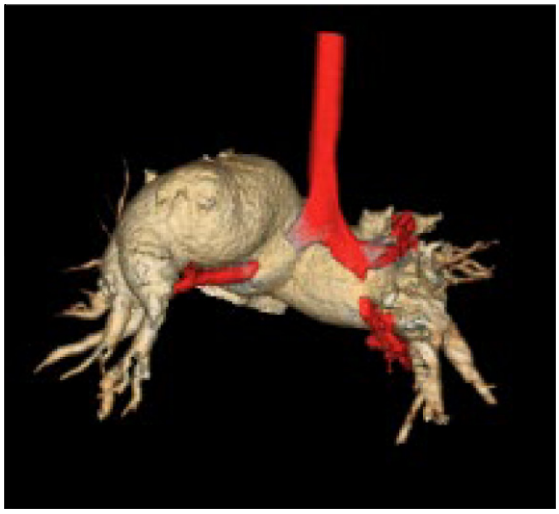

Figure 8 Posterior view of an infant with tetralogy of Fallot with absent pulmonary valve. Note complete compressive occlusion of the right and left main stem bronchi (red) by the overlying hugely dilated right and left branch pulmonary arteries.

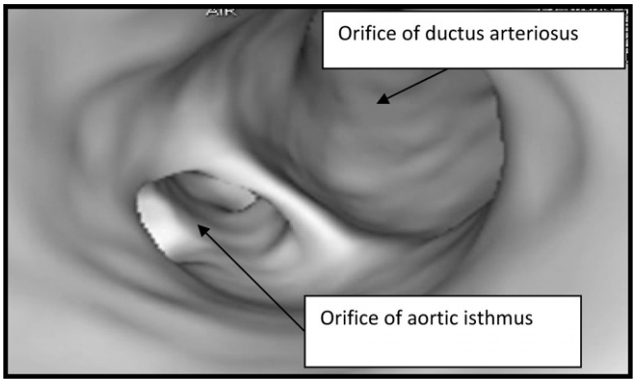

Figure 9 Example of infant arterioscopy showing the inner lumen of the distal aortic arch. Note the narrow isthmus representing coarctation and the orifice of a large patent ductus arteriosus.

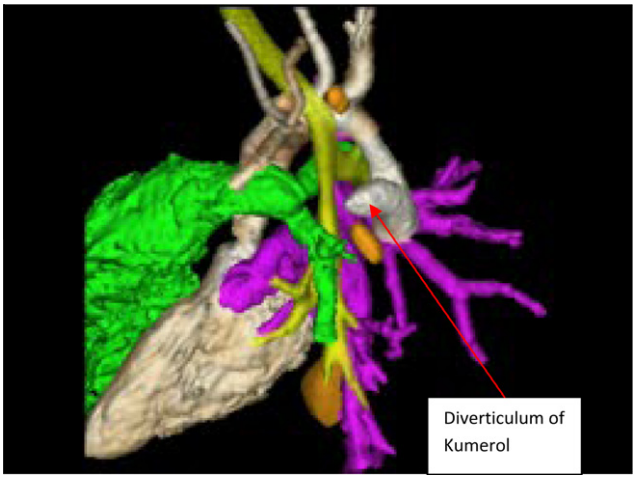

Figure 10 LV and aorta (natural); RV and PAs (green); trachea (yellow); esophagus (orange) in an infant with right aortic arch, vascular ring, and diverticulum of Kumerol.

Figure 11 Separate stents placed in the lumen of the left posterior and anterior branches of the left main pulmonary artery.

With MSCT a single bolus of 1–2 cc/kg administered via a peripheral vein suffices to opacify the entire cardiac structure leading to much smaller volumes of contrast compared to most DCC and ICC (Frush and Yoshizumi, 2006). Standard angiography exposes the patient, the physician, and the support staff to varying amounts of radiation. With MSCT, only the patient is exposed. MSCT exposes a volumetric area of interest and thus "sees" all pathology within that volume. Unexpected

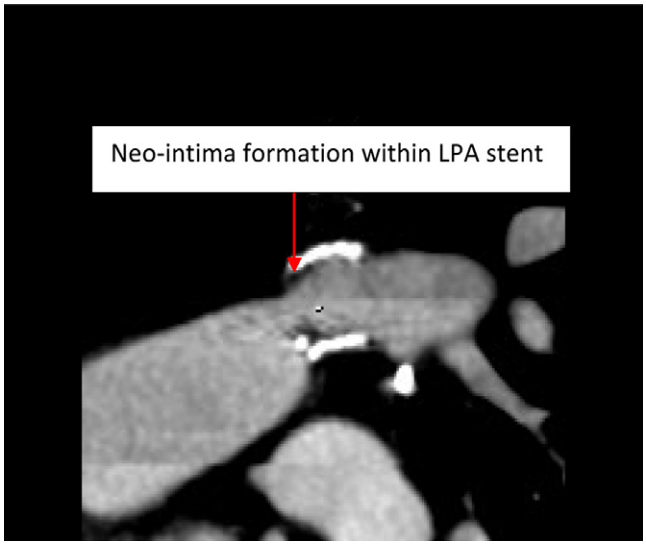

Figure 12 Neo-intima formation (red arrow) between the residual lumen and a previously placed LPA stent.

and unsuspected vascular pathology may be found that has the potential to change clinical management. In a review of 213 gated MSCT examinations conducted on 203 patients between August 2005 and October 2006, I found 78 instances of unexpected and unsuspected anomalies in 58 patients (Mathewson et al., 2006). Therapeutics were secondarily altered in 22%. Twenty-five percent involved coronary artery anomalies including anomalous origin of the left coronary from the pulmonary artery and anomalous origin of the right coronary artery from the left coronary cusp (Mathewson et al., 2008).

4. Over utilization and other drawbacks of MSCT

Very infrequently, MSCT may be over utilized. The most common reason relates to those situations where surgeons understand the utility of 3D imaging, have ready access to it, there is an absence of high quality 2DE acquired under properly sedated steady state conditions, and there is a lack of quality assurance controls for the use of 2DE, DCC, MRI, and MSCT.

For adult MSCT the effective RE in mSv may be estimated from the dose length product (DLP) which is directly obtained from the main frame software multiplied by an appropriate tissue constant related to the area of interest (Huda et al., 2008). Applying this same formula to infants and children may underestimate the RE and lead to the erroneous assumption that infants and children are receiving small doses. Pediatric RE is more complex to calculate compared to adults and requires accounting for the patient's weight, the RE received by an adult undergoing the identical procedure, pitch, gantry rotation speed, and the kV and mAs used (Huda and Ogden, 2008).

3D rendering is time consuming and labor intensive compared to viewing individual axial slices. Nevertheless, a non-gated study can be completed in less than 90 min. Retrospectively gated studies including volume calculations can take more than 180 min to fully analyze and render in 3D. But the value of the information obtained may be greater than that obtained from standard DCC, the performance and interpretation of which require roughly equal time.

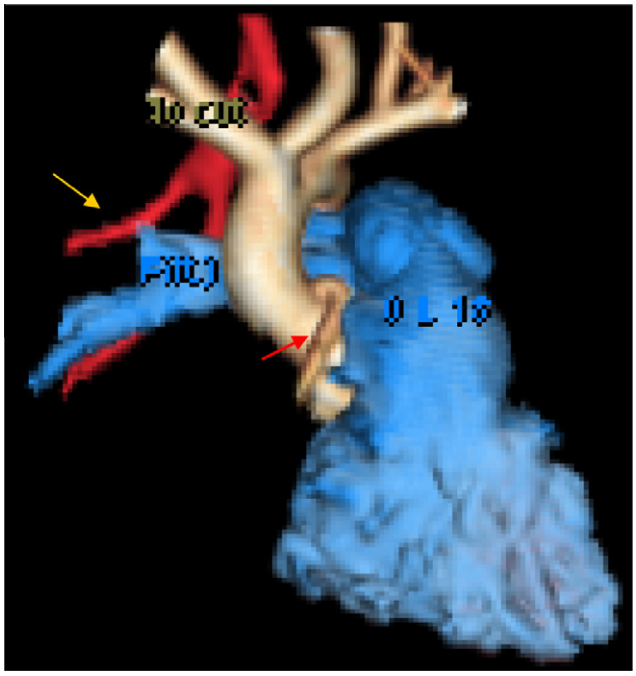

Figure 13 1.7 kg premie with HR 171bpm. Note pig bronchus (yellow arrow) and anomalous origin of the right coronary artery (red arrow) from the left anterior surface of the ascending aorta.

Another CT drawback is the requirement that there be no chest wall movement during image acquisition in order to reduce movement artifacts. Achieving effective apnea is simply part of the anesthesia learning curve and is soon mastered.

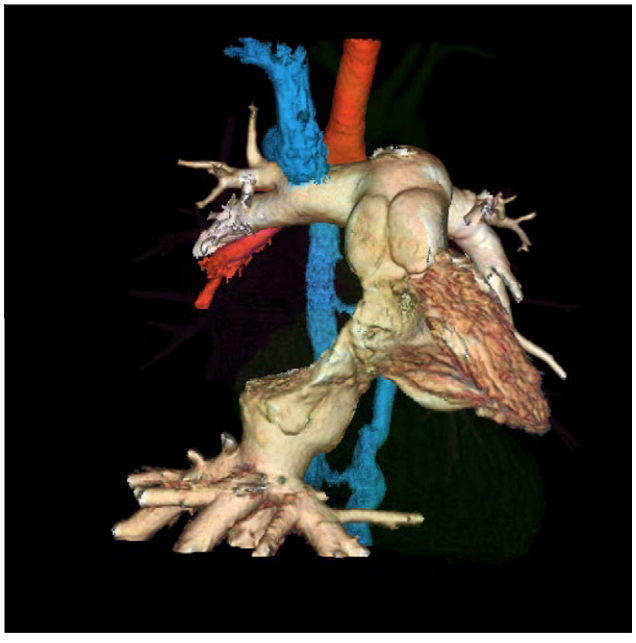

Figure 14 Adult with transposition and Hemi-mustard repair. Note the dilation of the proximal IVC and hepatic veins consistent with partial obstruction of the inferior baffle limb. The SVC is anatomosed to the RPA (Glenn anastomosis), and the azygous vein to the lower compartment is dilated suggestive of high upper compartment pressure. The LV gives rise to the main and branch pulmonary arteries (transposition).

The form of anesthesia needed for 5–15 s of complete apnea with no chest wall movement is very different from that required to support a patient for 30 min or more as is required for MRI, DCC or ICC. Often all that is needed is a short-acting muscle relaxant with or without pre-image-acquisition hyperventilation.

It is often incorrectly assumed that heart rates should be less than 80 bpm to achieve good CT imaging. In my experience, a regular HR is more important than the absolute HR, and neonatal and infant weight is not a factor. Fig. 13 shows a 1.7 kg premature infant sent for CT to determine the cause of unexplained RV enlargement and to rule out a pulmonary sling that was suggested by 2DE. This infant was found to have partial anomalous pulmonary venous connection of the right lung to the SVC/RA junction, no sling, anomalous origin of the right coronary artery from the left facing sinus (red arrow), and a right-sided pig bronchus (yellow arrow). Each finding was unexpected and unsuspected.

5. When is MSCT indicated and who might benefit?

When 2DE fails to provide information sufficient to make the desired clinical and/or surgical decisions, one should consider MSCT. This includes patients with unanswered questions who were not sedated for 2DE or adults with complex CHD (Fig. 14) with limited echo windows. It also includes patients with severe lung disease or when there is some form of isomerism with or without dextrocardia. Given the relatively comparable RE received during non-ECG and prospective ECG gating, MSCT is an attractive alternative to DCC or incomplete 2DE. Patients for whom CT is contemplated should be discussed at pre-procedure conferences to determine if the expressed need is due to an incomplete or poorly rendered echocardiogram that could be repeated, to decide collectively if another test is needed, and to determine the wisdom of exposing patients to radiation or long periods of general anesthesia. When CT is chosen, it should be used in the acquisition format that results in the least RE and is the most appropriate for the desired information.

6. Summary

MSCT is a valuable imaging tool that should be utilized more often for the diagnosis and management of CHD at all ages. Rejecting its use simply due to misconceptions about excessive RE is unwarranted given the uncontrolled and unknown RE during many DCC and ICC and electrophysiologic procedures. To reject MSCT because of ignorance of its full applicability is also no excuse. MSCT is especially attractive for adults with CHD and for pediatric patients with complex anatomy and physiology whose life expectancies and risk for radiation sequelae are different from those with less difficult CHD. An accurate 3D display of complex anatomy may be more beneficial to the patient's short-term surgical and/or clinical management and the resulting quality of life than multiple incompletely or poorly performed and interpreted 2DE, DCC, or MRI.

Finally, I believe it is essential for radiologists and cardiologists to work together to produce the most comprehensive diagnostic MSCT imaging set at the lowest radiation exposure possible. Radiologists know chest pathology well but are less well trained in congenital cardiac pathology. The reverse is true for cardiologists. Working together, patients will receive the maximum benefit at the lowest risk.

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James W. Mathewson
St. Joseph Hospital and Medical Center, Phoenix, Arizona, United States

National Guard Health Affairs Medical Center, Riyadh, Saudi Arabia
Tel.: +966 858 395 8652
E-mail address: jwmathewson@yahoo.com

Available online 21 July 2010